Published in final edited form as:

Otolaryngol Head Neck Surg. 2010 August ; 143(2): 269–273. doi:10.1016/j.otohns.2010.03.001.

Endolymphatic pseudohydrops of the cochlear apex

Andres Makarem, MD^{*}, Jose N. Fayad, MD^{**}, and Fred H. Linthicum Jr., MD^{***}

^{*}House Ear Institute. Los Angeles California

^{**}House Clinic, House Ear Institute, and University of Southern California Keck School of Medicine

^{***}House Ear Institute and University of Southern California Keck School of Medicine

Abstract

OBJECTIVE—To demonstrate that what appears to be endolymphatic hydrops of the apical scala media is normal anatomy.

STUDY DESIGN—Computer-generated three-dimensional reconstruction of the cochlear apex and tabulation of the number of cases with arched Reissner's membranes (pseudohydrops) vs. flat membranes.

SETTING—Temporal Bone Laboratory consisting of 809 documented pairs of temporal bones.

SUBJECTS AND METHODS—Archival temporal bone sections from 107 bones (65 patients) were used to determine the percent of arched (pseudohydrops) vs. flat Reissner's membranes. Two bones, one of each membrane shape, were randomly selected for computer-generated three-dimensional reconstructions showing the cochlear apical anatomy.

RESULTS—An arched Reissner's membrane was found in 48.6% of bones. In the cochlear apex, Reissner's membrane appears to be distended, simulating hydrops, due to its transition from a conical structure to a triangle bounded by the basilar membrane with the organ of Corti, the stria vascularis, and Reissner's membrane. Membrane findings were similar in both ears in 73.8% of the bilateral cases studied. There were no significant relationships between membrane type and clinical characteristics.

CONCLUSION—What appears to be endolymphatic hydrops of the cochlear apex is the transition area of the cochlear duct from a conical shape at the extreme apex to the triangular shape found in the rest of the cochlea. The appearance of distension is dependent upon the cochlear length and the level of the microscopic section.

Introduction

A curved configuration of Reissner's membrane in the cochlear apex, with the convexity into the perilymphatic space in man and experimental animals, is sometimes described as a

© 2010 American Academy of Otolaryngology Head and Neck Surgery Foundation, Inc. Published by Mosby, Inc. All rights reserved.

Corresponding author: Fred H. Linthicum Jr. MD House Ear Institute 2100 W. 3rd St. Los Angeles, CA 90057 213 252 7093 flinthicum@hei.org.

Publisher's Disclaimer: This is a PDF file of an unedited manuscript that has been accepted for publication. As a service to our customers we are providing this early version of the manuscript. The manuscript will undergo copyediting, typesetting, and review of the resulting proof before it is published in its final citable form. Please note that during the production process errors may be discovered which could affect the content, and all legal disclaimers that apply to the journal pertain.

Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

mild or early form of endolymphatic hydrops (Fig 1). Yamachita and Schuknecht described this finding and concluded that this apical “endolymphatic hydrops” had no clinical significance.¹ We have previously reported that this apparent hydrops is normal anatomy.² Since that time, one of us has been asked on several occasions to review submitted papers purporting to physiological significance of what appears to be cochlear apical hydrops in man and some experimental animals. We therefore decided that it was time for another analysis of this issue to demonstrate its frequency in patients other than those with a clinical diagnosis related to cochlear hydrops.

Methods

This research involved temporal bone sections (specimens) of deceased individuals, not living individuals, therefore it does not fall under OHRP’s definition of research involving human subjects (45 CFR 46.101(b)(4)). As a result, this study is exempted from IRB approval.

The temporal bones in our institution’s bone bank were decalcified in EDTA for 8-12 months, embedded in celloidin, and sectioned at intervals of 20 μ m. The sections were then put on slides and analyzed under light microscopy. A total of 122 sequential cases (244 temporal bones) were randomly selected for the study. Bones with true cochlear hydrops (clinical diagnosis of cochlear hydrops or Ménière’s disease) and those with enough artifact to prevent accurate examination were excluded from the final analysis, leaving a total of 165 temporal bones. Of these, 107 bones from 65 patients had clinical records available and were included in the study. These included both ears of 42 patients and 23 unilateral cases. The clinical characteristics of the patients are summarized in Table 1, including demographics, age at death, hearing at last hearing test and diagnosis. We also examined the inner ears of other species for this study, including a guinea pig, dog, cat, mouse, rabbit, and baboon (Fig 1).

Two bones were randomly selected for virtual three-dimensional reconstruction with the Amira 4.0 software ®; (Mercury Computer systems, Inc, Chelsford, MA, USA), the first with an arched apical Reissner’s membrane and another with a straight membrane. Every other section was stained with H&E and placed on slides. Each stained section was digitally photographed under a 20 X magnification and uploaded into Amira 4.0. The images were correctly oriented using the AlignSlices tool and turned to monochannel for labeling. The structures selected for labeling were Reissner’s membrane, perilymph, endolymph, stria vascularis, spiral ligament, organ of Corti and bone. The next step was to create virtual slides between the real ones using InterpolateLabels for higher accuracy in the reconstruction. Next, the program was able to build the three-dimensional object with the SurfaceGen tool, and the labels were retouched manually to avoid mistakes in the object’s anatomy. This reconstruction could be rotated in any direction in real time and allowed us to make a very detailed evaluation of the conical shape of the cochlear duct origin in the cochlear apex.

Analysis was performed using the LinearProbe tool, which measures linear distances between two points in space. SurfaceCut was used to find the locations where Reissner’s membrane first looked hydropic and then straight. One probe was positioned in each of these points and the linear distance (perpendicular to the cutting plane) measured.

Variations in length and size of the cochlea have been previously described.³⁻⁶ In our collection there are eight temporal bones which were sectioned in a coronal plane and demonstrate this variation. These cochleae demonstrate how different orientations and lengths might change the appearance of Reissner’s membrane during sectioning. Therefore,

we rotated the reconstructed apical segment to mimic the different orientations (Fig 2). Surface Cut has a rotation option which was used to ensure that the orientation of the cutting plane was axial after the structure's rotation. Once these changes were made, the same analysis was done on the rotated cochleae.

Data Analysis

In addition to descriptive statistics, inferential statistical testing was performed to determine any relationships between membrane type and clinical characteristics. Chi-Square analysis was used for categorical variables (sex, right ear membrane type vs left ear membrane type, diagnosis) and independent group t-tests for continuous variables (age, pure-tone average hearing threshold). Criterion for statistical significance was set at $p \leq .05$, two-tailed.

Results

Over all ears, 48.6% had an arched RM. Of the bilateral cases, the membrane was arched in both ears in 38.1%, flat in both ears in 35.7% (i.e., the same on both sides in 73.8%), and different in the two ears in the remaining 26.2%. This was a statistically significant relationship between membrane status in the two ears ($p \leq .002$).

There was no significant relationship between membrane type and sex; 49.3% of females and 46.9% of males had an arched RM. There also appears to be no relationship between diagnosis and membrane type, although the number of cases with any one diagnosis is too small for statistical analysis. Of those with otosclerosis, 44.7% had an arched RM as did 52.9% of those with sensorineural hereditary loss and 56.3% of those with presbycusis. There was no significant difference in PTA or age between those with an arched or a flat membrane.

Apical reconstruction of the cochlea with an arched Reissner's membrane shows that the cochlear apex is entirely occupied by the endolymphatic duct (Fig 3A). At this level, Reissner's membrane (adjacent to the cochlear wall) and the stria vascularis form a blind sac in the shape of a blunted cone at the apex. A few microns away from the cochlear apex, the perilymph begins to appear as Reissner's membrane begins to detach from the cochlear wall (Fig 3B). Finally, the cochlear duct reaches its characteristic triangular shape, however the membrane is arched and appears slightly hydropic (Fig 3C, Fig. 4).

Apical reconstruction of the cochlea with a flat Reissner's membrane shows a different cochlear orientation. The apex is laterally oriented (Fig. 5), which makes it parallel to the sectioning plane. After removing all structures, using the computer software, except for Reissner's membrane, stria vascularis, organ of Corti and basilar membrane, the apical portion of the scala media appeared to be the same as the previous reconstruction: Reissner's membrane in the shape of a rounded cone. When the cochlea was rotated to match the orientation of the previous one with an arched Reissner's membrane and the sectioning plane was correctly aligned, Reissner's membrane was shown to behave the same.

The initial reconstructed cochlea has the most common orientation found on temporal bones (Fig 2). The distance between the apex and the straight Reissner's membrane was found to be 789 microns. This means that, if the scala media always expands into a blind sac at the cochlear apex, Reissner's membrane would look hydropic in 3 or 4 stained sections (classically, every stained section is 200 microns apart).

Once the cochlea was rotated to resemble the other orientations, the results began to vary. In one orientation, it was not possible to see the membrane become straight from its hydropic appearance in the upper turn. In others, the sectioning angle prevents Reissner's membrane

from seeming hydropic in any section, so the membrane appears straight as soon as it is visible (Fig 5). The distance between the apex and the straight section is 335 microns, which means that in one section the membrane would look hydropic.

Discussion

Our findings demonstrate that in the cochlear apex, Reissner's membrane frequently appears to be distended, simulating hydrops. This is due to its transition from a conical structure to a triangle bounded by the basilar membrane with the organ of Corti, the stria vascularis, and Reissner's membrane. The cochlear duct in the apex of the cochlea appears on three-dimensional reconstruction to be a blunted cone with the tip occurring in different relationships to the rest of the cochlea, and thus the plane of sectioning, depending on the length of the cochlea.

We found a 48.6% rate of arched Reissner's membranes in our temporal bone bank samples. Yamahsirta and Schucknecht also described this phenomenon many years ago, although they found it in only 10% of cases.¹ They categorized the degree of hydrops as mild, moderate, or pronounced depending on the degree of Reissner's membrane displacement; but they note that the judgment of the degree of apparent hydrops is an arbitrary one. In this study, we defined any evidence of arching of the membrane into the perilymph space as hydrops. We also had the opportunity to use new technology to provide three-dimensional reconstructions that help explain the arched appearance of the membrane in many temporal bones.

Pseudohydrops is found in both normal temporal bones and temporal bones with unrelated pathology. Thus far, pseudohydrops has been an incidental finding with no known explanation or significance. We found no significant relationships of membrane type to clinical characteristics. More importantly, bones from patients with any of the clinical symptoms diagnostic for cochlear hydrops (i.e., Ménière's disease or cochlear hydrops) were excluded from study. In addition, it is a feature that isn't exclusive to humans. Our findings suggest that the basic reason for the different presentations of Reissner's membrane in the apical part of the cochlea is anatomical variability. In the most common cochlear orientation during sectioning, the membrane appears arched and hydropic in the apex. In certain orientation variants, which include cochleae with different lengths, the sectioning angle prevents us from seeing the true anatomy of the apex, and Reissner's membrane appears as a linear structure from beginning to end.

Conclusion

In the cochlear apex, the scala media begins as a conically-shaped blind sac, and Reissner's membrane surrounds the scala almost in its entirety. As it progresses basally, it adapts to the well-known triangular shape. During this transformation, the arched membrane gives the impression of being hydropic, but this is only part of its anatomy, and thus should be considered normal.

Acknowledgments

The authors thank Karen I. Berliner, PhD for data analysis and editorial help, and Diana Cohen and Ana Cortez for preparation of the slides used in the 3D reconstructions.

Supported by: NIDCD of NIH, #1R24DC008625-01 and House Ear Institute

References

1. Yamashita T, Schuknecht HF. Apical endolymphatic hydrops. *Arch Otolaryngol* 1982;108:463–6. [PubMed: 7103821]
2. Lau SK, Linthicum FH Jr. Apparent apical endolymphatic hydrops: computer-aided three-dimensional reconstruction and histologic study of the apical turn of the cochlear duct. *Am J Otol* 1993;14:79–81. [PubMed: 8424482]
3. Ulehlová L, Voldrich L, Janisch R. Correlative study of sensory cell density and cochlear length in humans. *Hear Res* 1987;28:149–51. [PubMed: 3654386]
4. Kawano A, Seldon HL, Clark GM. Computer-aided three-dimensional reconstruction in human cochlear maps: measurement of the lengths of organ of Corti, outer wall, inner wall, and Rosenthal's canal. *Ann Otol Rhinol Laryngol* 1996;105:701–9. [PubMed: 8800056]
5. Sato H, Sando I, Takahashi H. Sexual dimorphism and development of the human cochlea. Computer 3-D measurement. *Acta Otolaryngol* 1991;111:1037–40. [PubMed: 1763623]
6. Tian Q, Linthicum FH Jr, Fayad JN. Human cochleae with three turns: an unreported malformation. *Laryngoscope* 2006;116:800–3. [PubMed: 16652091]

Figure 1.
(A) Midmodiolar section of the human cochlea showing arched Reissner's membrane of the apical segment (arrow). X20, Hematoxylin and eosin (H&E). (B) Arched Reissner's membrane in rabbit and (C) guinea pig, X 40 (H&E)

Figure 2.

A demonstration of the varying orientation of the apical segment in two of the eight temporal bones cut in the coronal plane. The yellow indicates the organ of Corti on the white basilar membrane. X10

Figure 3.

(A) The cochlear duct (a) occupies the entire area of the most apical aspect of the cochlea. (b) The scala media in the middle cochlear segment. (B) A few microns away from the apex, Reissner's membrane begins to separate from the cochlear wall (arrow). (a) Indicates endolymphatic spaces. (C) Final triangular configuration of the scala media, but still showing some slight outward bulging of Reissner's membrane. (H&E X 40)

Figure 4. Computer-assisted three-dimensional reconstruction of the cochlear apex showing the conical configuration of the scala media apex. green = Reissner's membrane, red = stria vascularis, yellow = organ of Corti, white = basilar membrane and osseous spiral lamina, grayish blue = boundaries of perilymph spaces. X 20

Figure 5. Computer-aided reconstruction of a routine sectioned cochlear apex in which Reissner's membrane appears flat due to the variation in cochlear lengths. However, at the apex of the cochlea, the cochlear duct is a blunted cone. light blue = perilymph, red = stria vascularis, green = Reissner's membrane, dark blue = endolymph, yellow = organ of Corti, white = osseous spiral lamina. X20

Table 1

Characteristics of 65 patients (107 ears) whose temporal bones are included in this study

Ear: R/L/bilateral	13/10/42
Sex: M/F	32.3% / 67.7% [by ear: 29.9% / 70.1%]
Age: Mean (SD) years	78.6 (10.1) [by ear: 79.2 (10.0)]
PTA: Mean (SD)	47.4 (22.7) dB [by ear]
Diagnosis [by ear]	
Otosclerosis	43.9%
Presbycusis	15.0%
SN* hereditary	15.9%
Cochlear otosclerosis	6.5%
Chronic otitis media	3.7%
Normal	2.8%
Tumors**	2.7%
Other***	9.5%

* SN=sensorineural

** Tumors= one each of glomus jugulare, meningioma, vestibular schwannoma

*** Other=Bell's palsy, postural vertigo, SN cause undetermined/noise-induced/postop stapes/sudden, CNS vertigo.